RESTRICTIVE LUNG DISEASE

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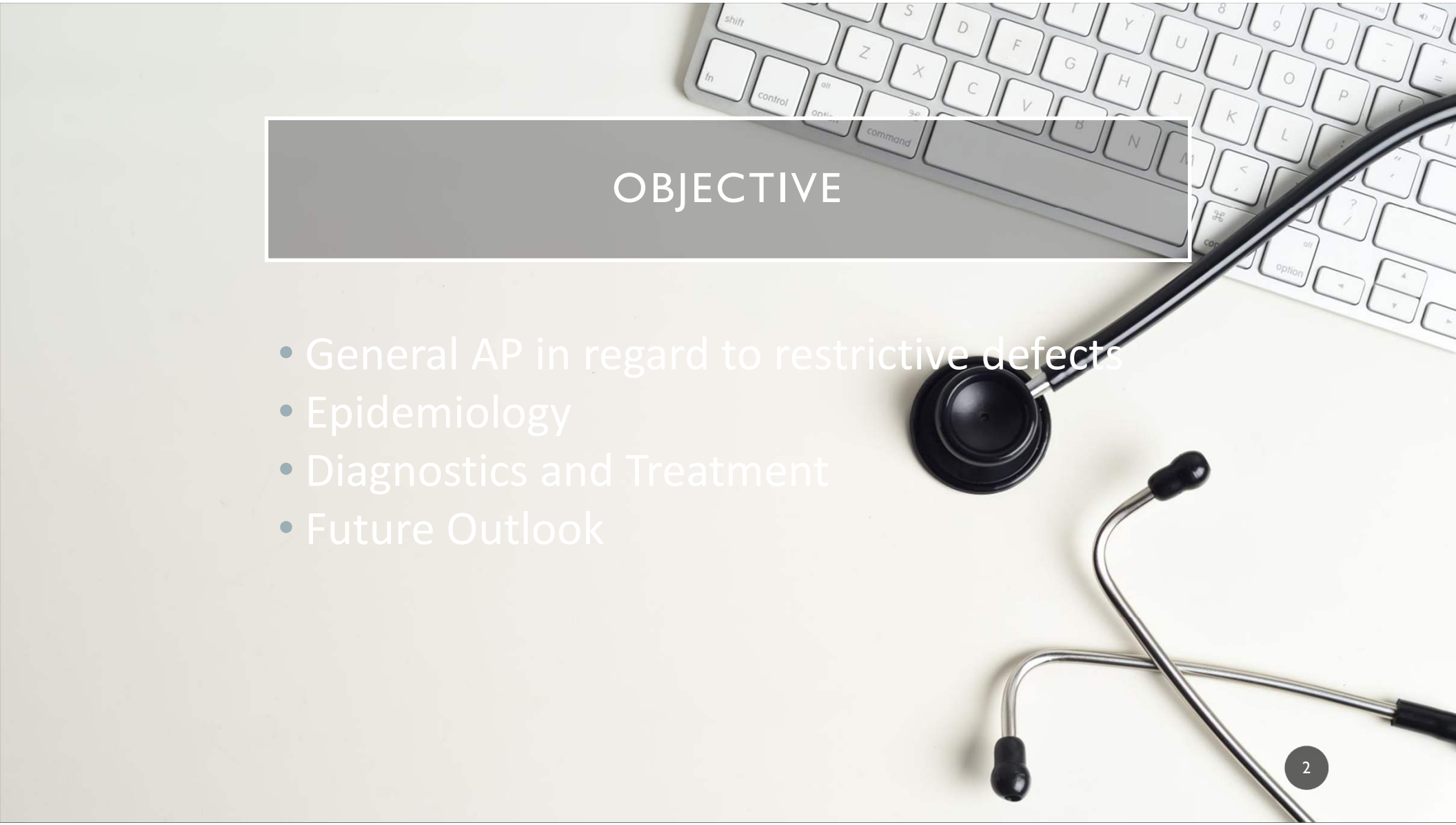
PULMONARY DISEASE NAVIGATOR AND RESPIRATORY EDUCATOR

OBJECTIVE

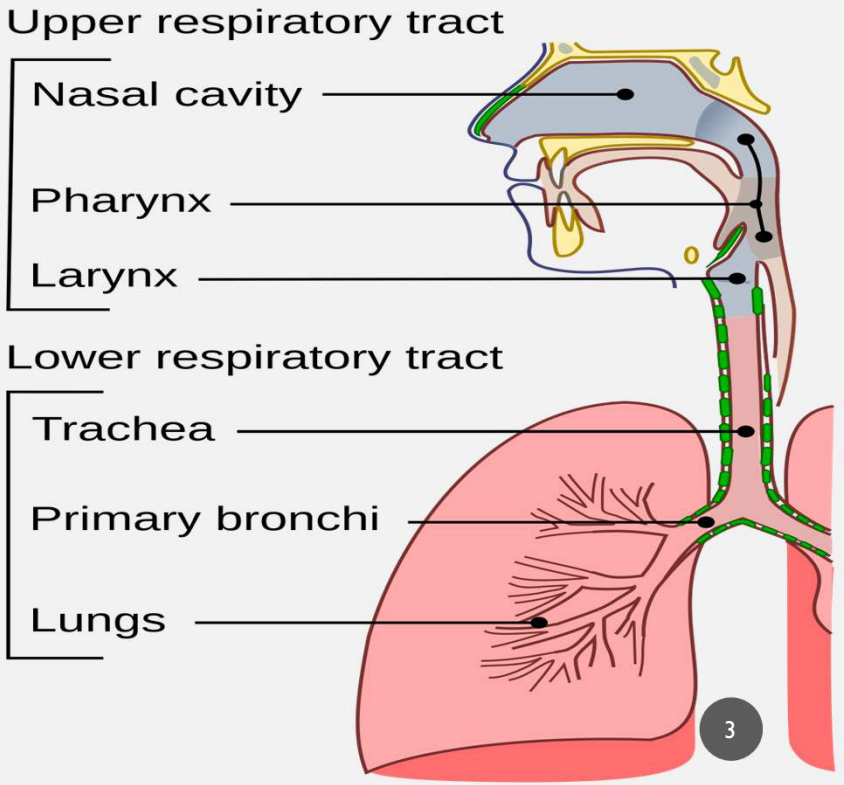
- General AP in regard to restrictive defects
- Epidemiology
- Diagnostics and Treatment
- Future Outlook



GENERAL LUNG AP

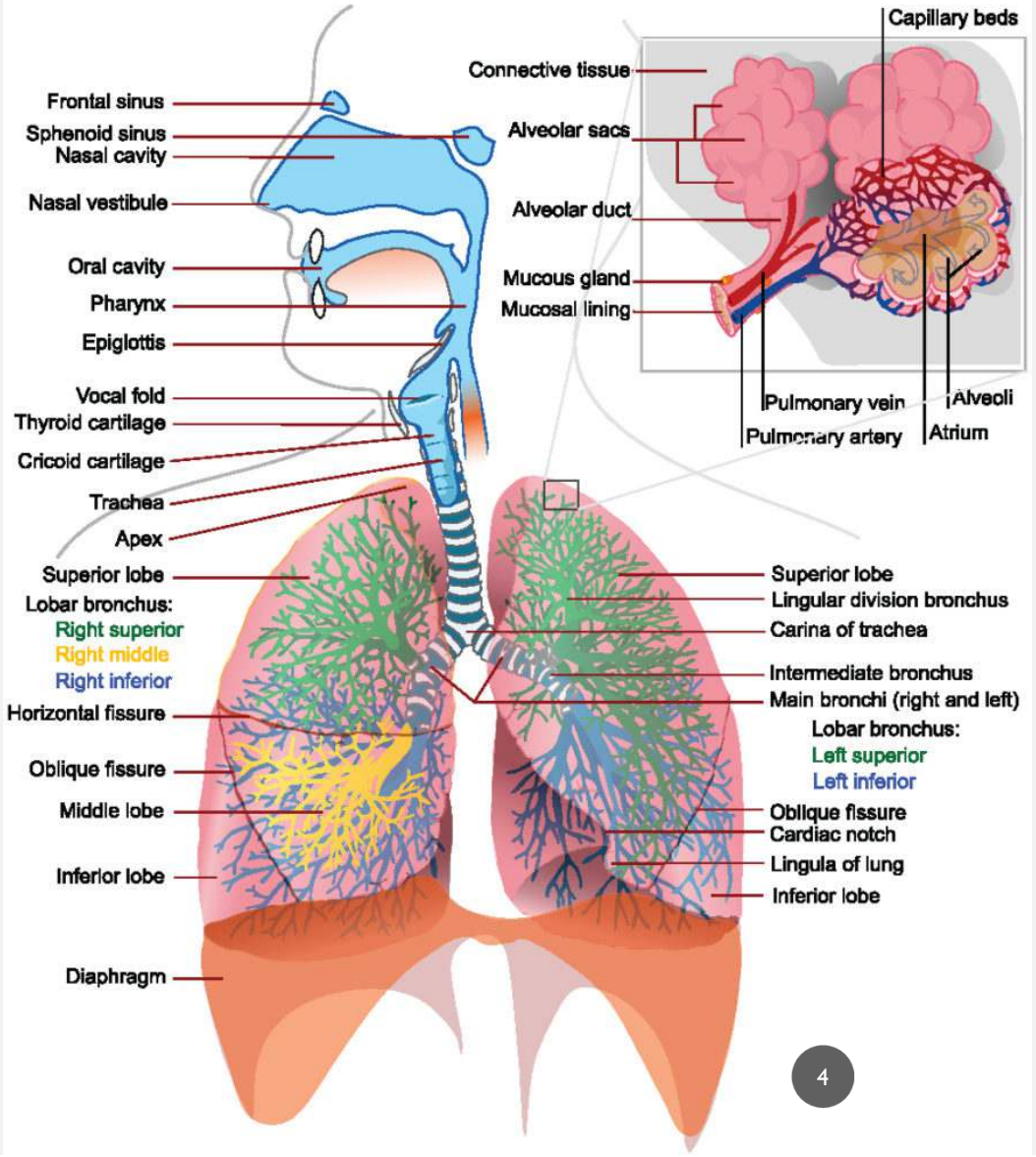
- Let's look at this like the magic school bus – were going start kind of big and bus it all the way down
- In the same way – it'll start quite easy and quickly get somewhat technical
- It is most important to know the body before sin and after – rather – as it should be vs how it sometimes comes to be.
- Restrictive lung disease comes in many different flavors

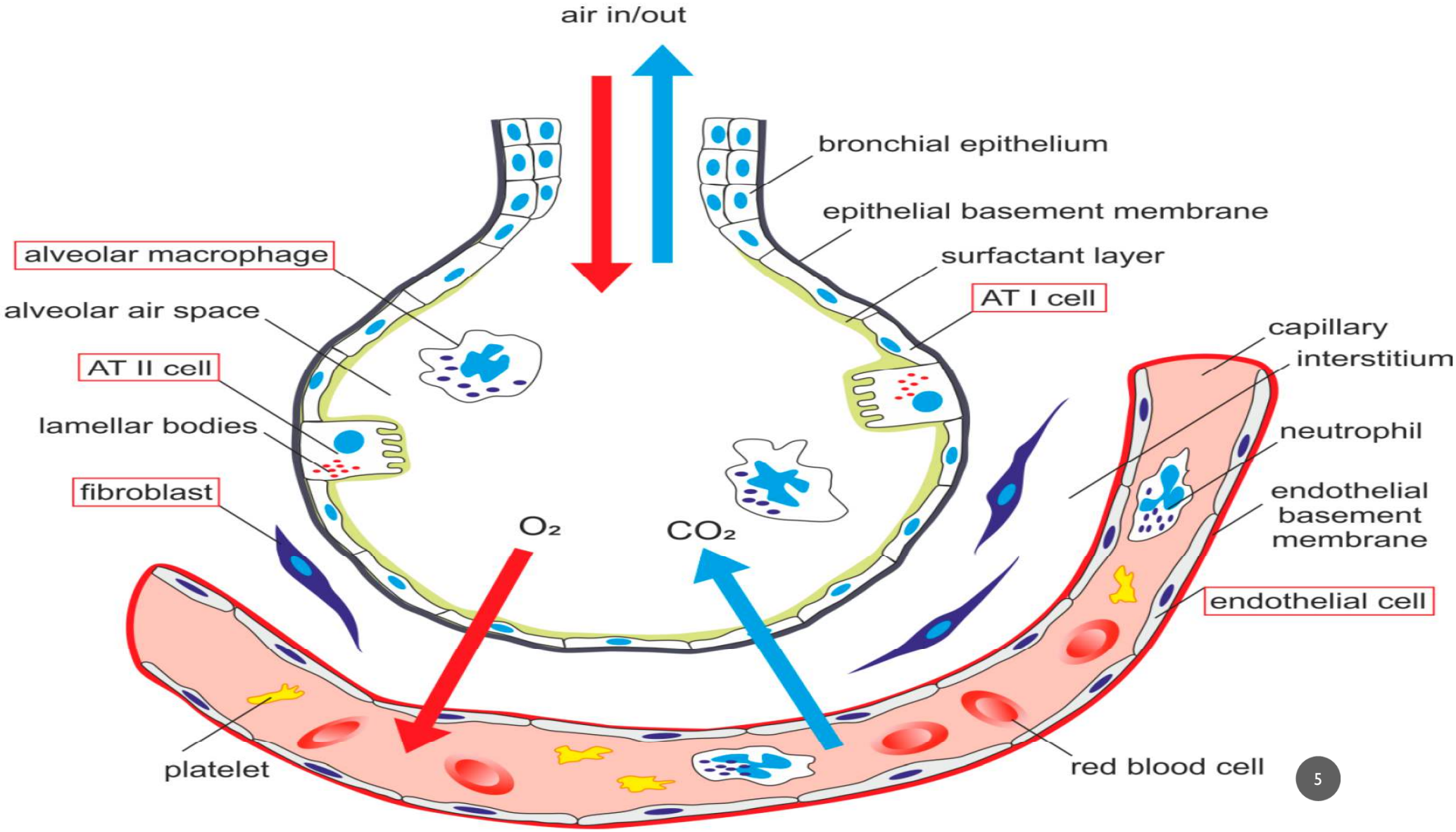
- Well, lets follow an oxygen molecule and start right past the larynx, shall we?
- At this point we have bypassed all the upper airway and all of its quirks and have made our way into the lower air way



GENERAL LUNG AP

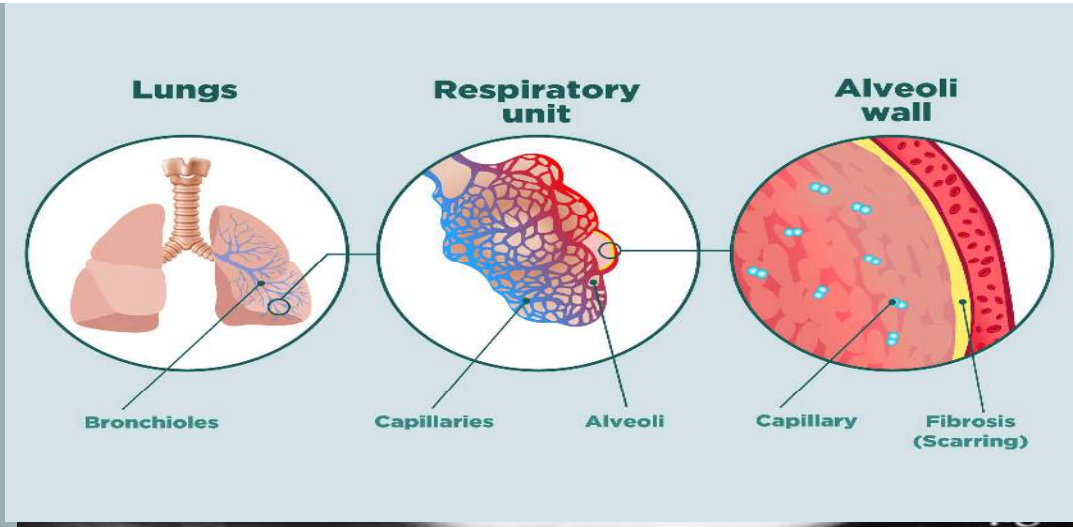
- The conducting airway
  - Trachea
  - Mainstem bronchi
  - Lobar bronchi
  - Segmented and sub-segmented
  - Terminal bronchi
- The parenchyma
  - Respiratory bronchi
  - Alveolar ducts
  - Alveolar sacs

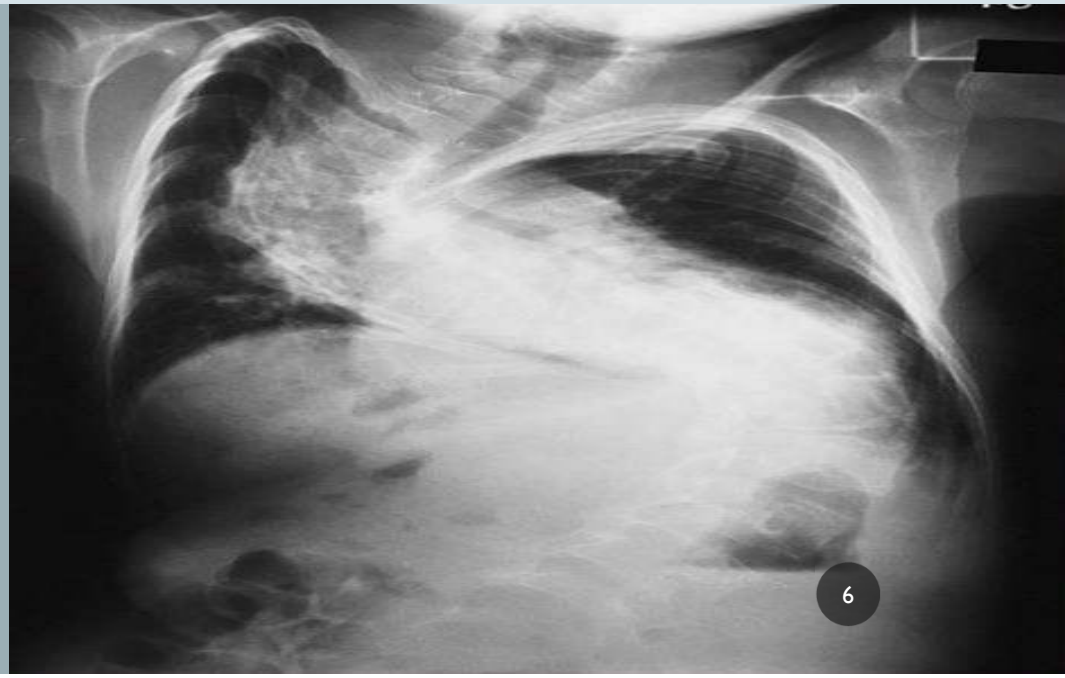



air in/out
bronchial epithelium
epithelial basement membrane
surfactant layer
AT I cell
alveolar macrophage
alveolar air space
AT II cell
lamellar bodies
fibroblast
capillary
interstitium
neutrophil
endothelial basement membrane
endothelial cell
O2
CO2
platelet
red blood cell

WHAT IS RLD

- Restrictive lung disease as a set of pulmonary disorders defined by restrictive patterns on spirometry…and lung volumes.
- Restrictive lung disease comes in many different flavors >200
- Think PAINT when thinking of types
  - Pleural
  - Alveolar
  - Interstitial
  - Neuromuscular
  - Thoracic cage





IMPORTANCE, PREVALENCE, AND CAUSES

- Importance of diagnosis and treatment
  - Early intervention – timely diagnosis and early treatment
  - Accurate treatment - Different types of restrictive lung diseases require different management approaches, and an accurate diagnosis ensures the most effective treatment is administered.
  - Symptom Management: Diagnosis enables healthcare providers to address the specific symptoms experienced by individuals with restrictive lung disease.

- **Prevalence**: around 3-6/100,000
- The prevalence of restrictive conditions increases from 2.7 cases per 100,000 persons in individuals aged 35-44 years to over 175 cases per 100,000 in those older than 75 years.
- African Americans- Compared to Whites (10.9 cases per 100,000), the prevalence in this group is 35.5 cases per 100,000 persons.
- It was found that restrictive patterns on spirometry were found to fall from 7.2% to 5.4% from 1988-1994 and from 2007-2010, which may be related to increased occupational safety precautions and falling rates of cigarette smoking.

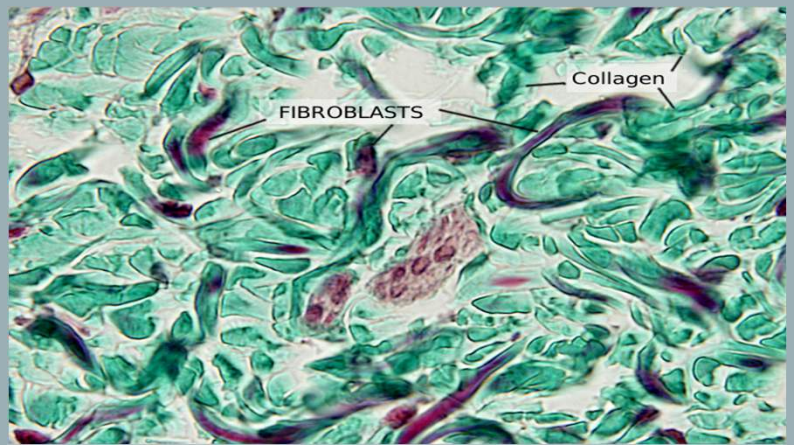
IMPORTANCE, PREVALENCE, AND CAUSES

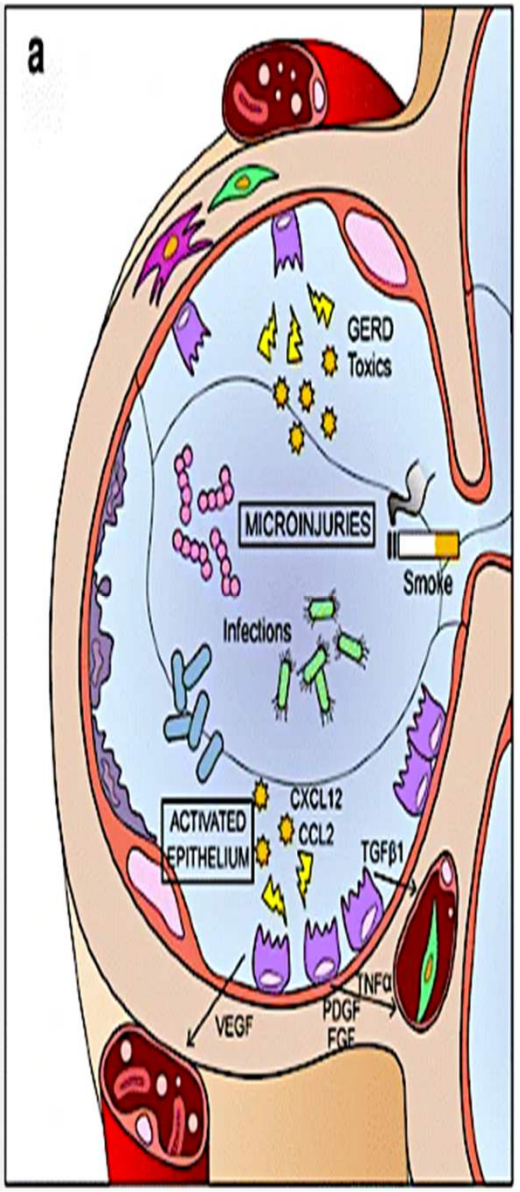
- Intrinsic Causes - restriction due to inflammatory processes within the lung tissue by diseases categorized under interstitial lung diseases.
  - IPF, Pneumoconiosis, Sarcoidosis and more
- Extrinsic Causes
  - Everything outside of the parenchyma
  - Boney, pleural, neuromuscular abnormalities
  - Obesity – complicates everything
  - Fluid or pressure build up in another part of the body such as acites

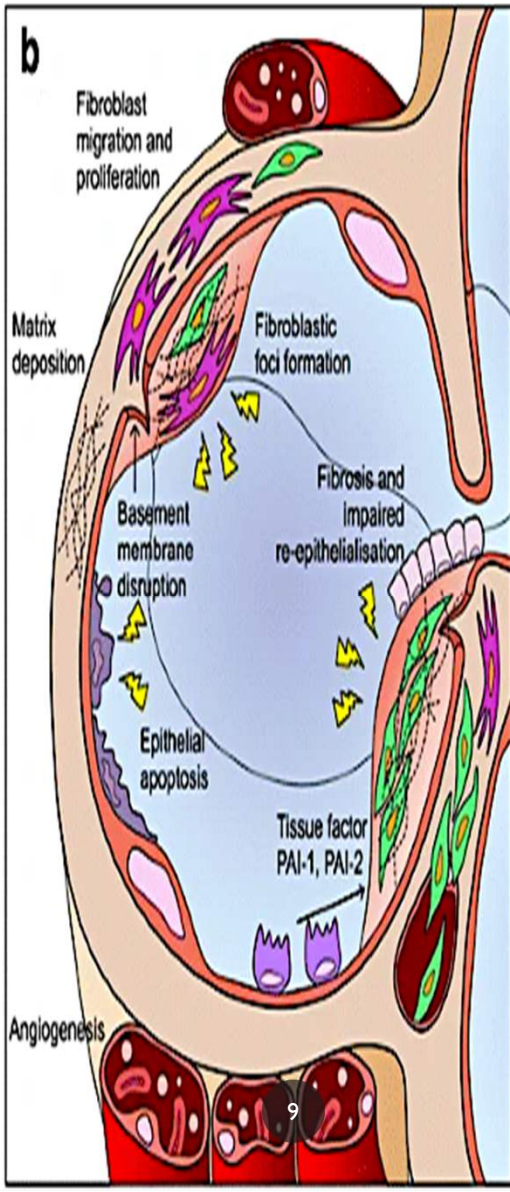
- Lets not forget occupational hazards

GENERAL RLD PATHOLOGY

- Most commonly associated with chronic inflammation of the lung parenchyma – this leads to over action of fibroblasts and secondarily collagen in the interstitium – which leads to fibrosis of the ECM.
- In many cases it goes much further than the scope of this lecture.



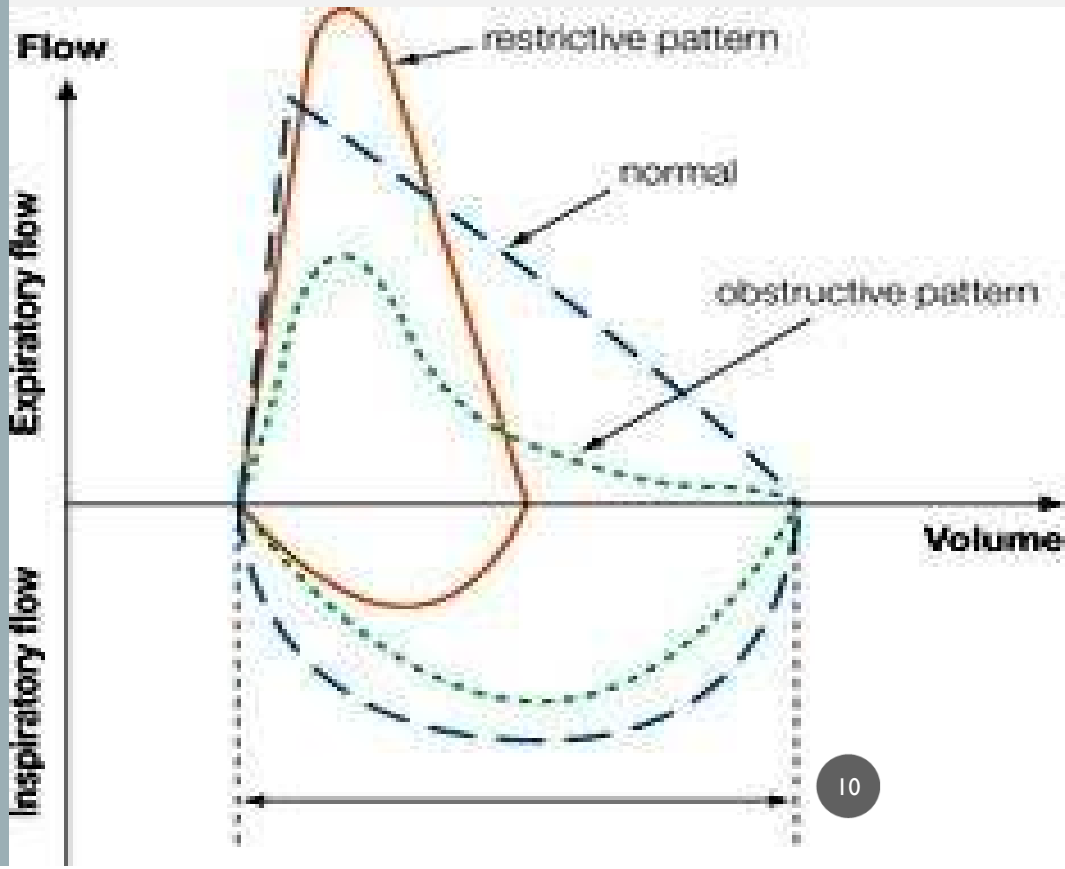




DIAGNOSIS

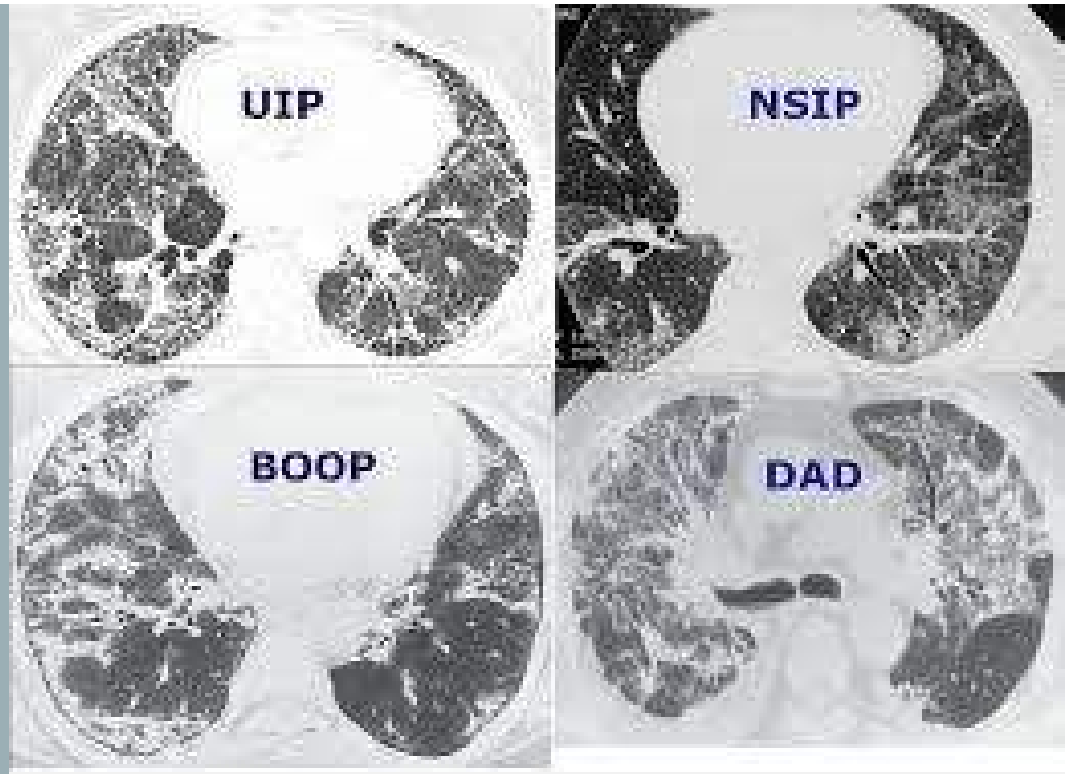
- Medical History
  - Duration or onset of illness
  - Family, occupational, and smoking hx
  - Prior medication use
  - Signs and symptoms
- PFT – Big thing here are VOLUMES
  - TLC < 80%
  - 70-80% - mild
  - 60-70% - moderate
  - 50-60% - moderate severe
  - < 50% - severe

- Possibly decreased FVC
- FEV1 is possibly normal, and in some cases, above normal.
- Preserved FEV1/FVC ratio or increased



DIAGNOSIS

- Imaging
  - High res. CT is the big one
- Other
  - Inflammatory markers
  - Antibodies
- Lung Biopsy (VATS)
  - If there is still no diagnosis after clinical evaluation, CT imaging and additional diagnostic tests, the provider may decide that a lung biopsy is needed.



Bronchiolitis Obliterans Organizing Pneumonia
Diffuse Alveolar Damage
Nonspecific interstitial pneumonia
Usual interstitial pneumonia

TREATMENT

- As always, we want to treat the underlying cause - in this case, if at all possible. Manage other comorbidities according to their established guidelines
- Pulmonary rehabilitation programs combine exercise, breathing techniques, and education to improve lung function and quality of life.
- Vaccines – cmon, right?
- Life style changes such as smoking cessation
- Surgical intervention such as transplant

- Medications
  - Bronchodilators: These medications help relax and widen the airways, making it easier to breathe.
  - Anti-inflammatory drugs: Corticosteroids or other anti-inflammatory medications may be prescribed to reduce lung inflammation and manage symptoms.
  - Immunosuppressive drugs: If the restrictive lung disease is caused by an autoimmune condition, medications that suppress the immune system may be used to reduce inflammation and preserve lung function.
  - Oxygen Therapy: Supplemental oxygen may be prescribed if oxygen levels in the blood are low, helping improve oxygenation and reducing shortness of breath.

TREATMENT

- Pirfenidone (Esbriet):
- Mechanism of Action: Pirfenidone is an antifibrotic medication that works by reducing the production of fibroblasts and inhibiting the synthesis of certain growth factors involved in the progression of fibrosis.
- Administration: Pirfenidone is available in oral tablet form and is typically taken three times a day with meals.

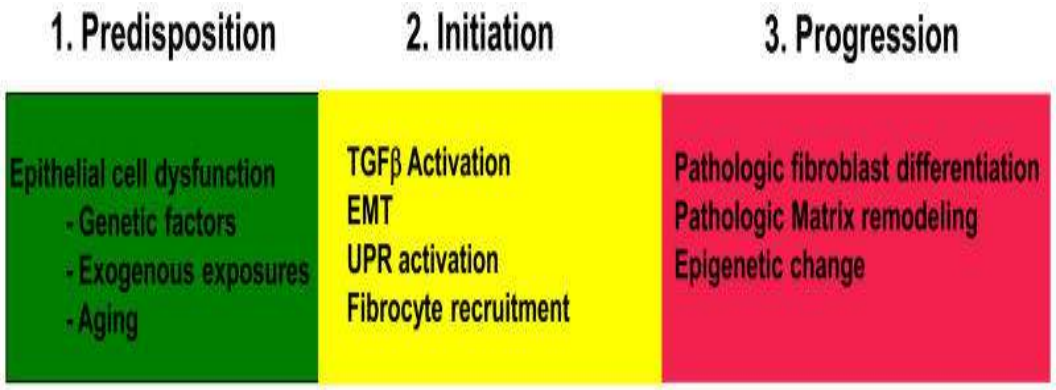
- Mechanism of Action: Nintedanib is a tyrosine kinase inhibitor that targets multiple growth factors involved in the development of fibrosis. It inhibits the signaling pathways that promote the proliferation, migration, and activation of fibroblasts.
- Administration: Nintedanib is available in oral capsule form and is typically taken twice daily with food.
- Both have been found to improve lung function, exercise capacity, and quality of life decrease progression of decline and exacerbations.

THE RT ROLE

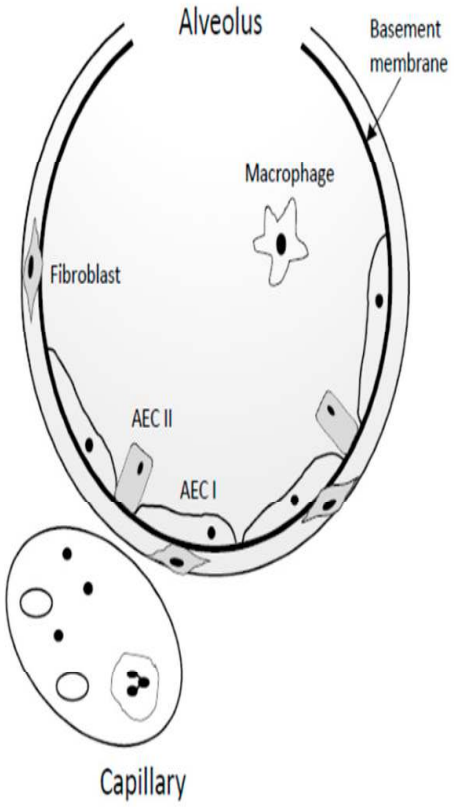
- Assessing lung function
  - Many RTs work in pulmonary function or are able to interpret PFT results
  - The same can be said for a vent as well
- Oxygen therapy
  - Low flow – high flow and everything between and beyond
- Vent support
  - Be aware of plat pressures and the like
    - These lungs are stiff, low compliance
- Airway clearance
- Medication administration
- Pulmonary rehabilitation - design and implement exercise programs tailored to individual patients' needs, monitor exercise tolerance, and provide education on breathing techniques, energy conservation, and self-management strategies.
- Collaborative care - work closely with other healthcare professionals, such as pulmonologists, nurses, and physical therapists, as part of an interdisciplinary team.

IPF

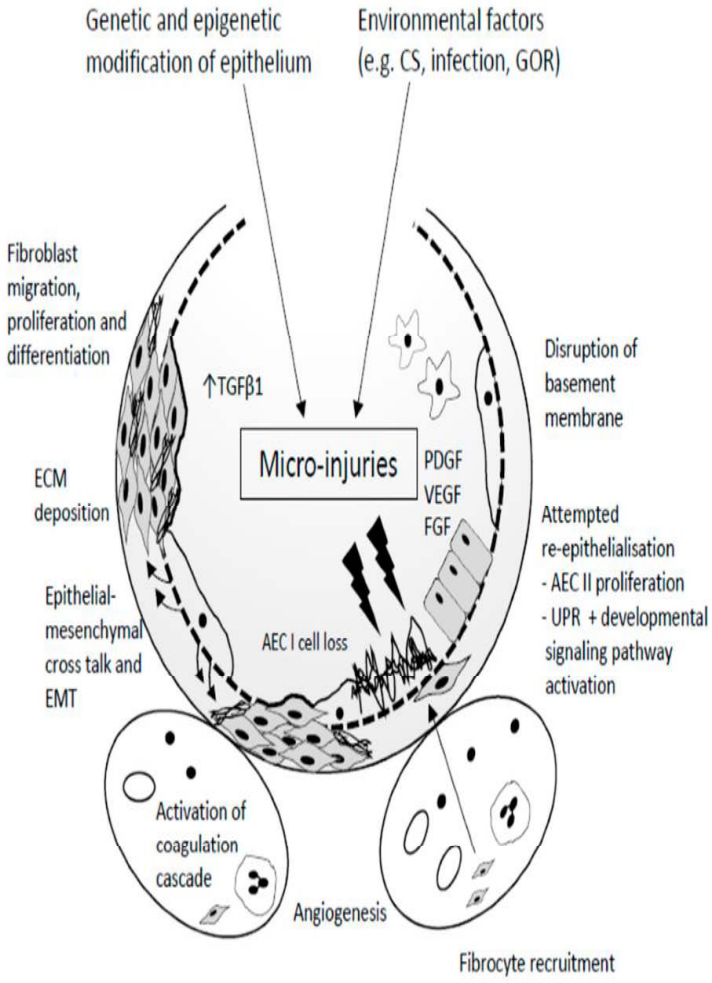
- Most common type of fibrotic lung disease
- White men between the ages of 50-70 are at most risk for IPF complications
- White, Black, and Hispanic, respectively.
- Common comorbidities include COPD, Hypertension, and Diabetes – shocker, right?
- According to NIH – roughly 30-40k new people are dx each year, however, ranges are all over the place
- Can be hereditary



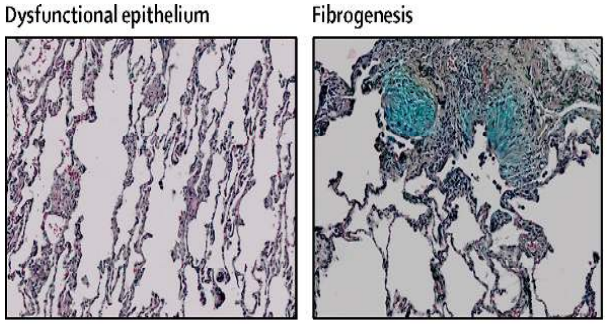
- Occupational hazards are a certain highway to RLD or IPF
- Recurrent exposure to toxins, chemicals, dust, etc., can scar the lungs over time
- #5 OSHA violation in 2019-2020?
- Progressive worsening of dyspnea (especially during exercise, cough, tiredness, in later stages – clubbing, gradual weight loss



Normal



Idiopathic Pulmonary Fibrosis

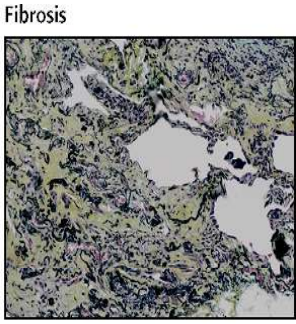


Dysfunctional epithelium

- Genetic susceptibility
- Ageing
- Recurrent microinjury

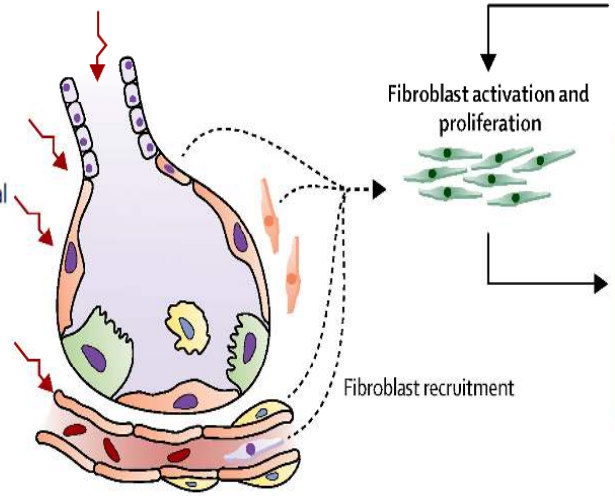
Fibrogenesis

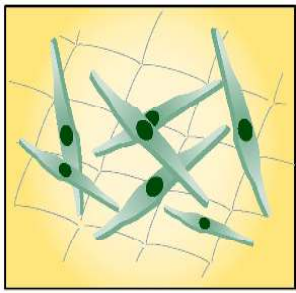
- Epithelial cell apoptosis and senescence



Fibrosis

- Extracellular matrix expansion
- Altered extracellular matrix composition
- Altered extracellular matrix biomechanics
- Deficient fibroblast apoptosis
- Alveolar collapse

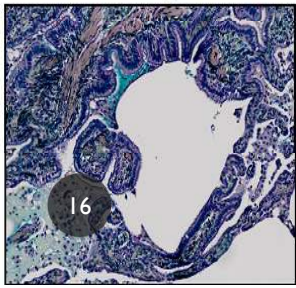




- Activation of epithelial cells
- Basement membrane disruption
- Dysregulated signalling
- Immune activation

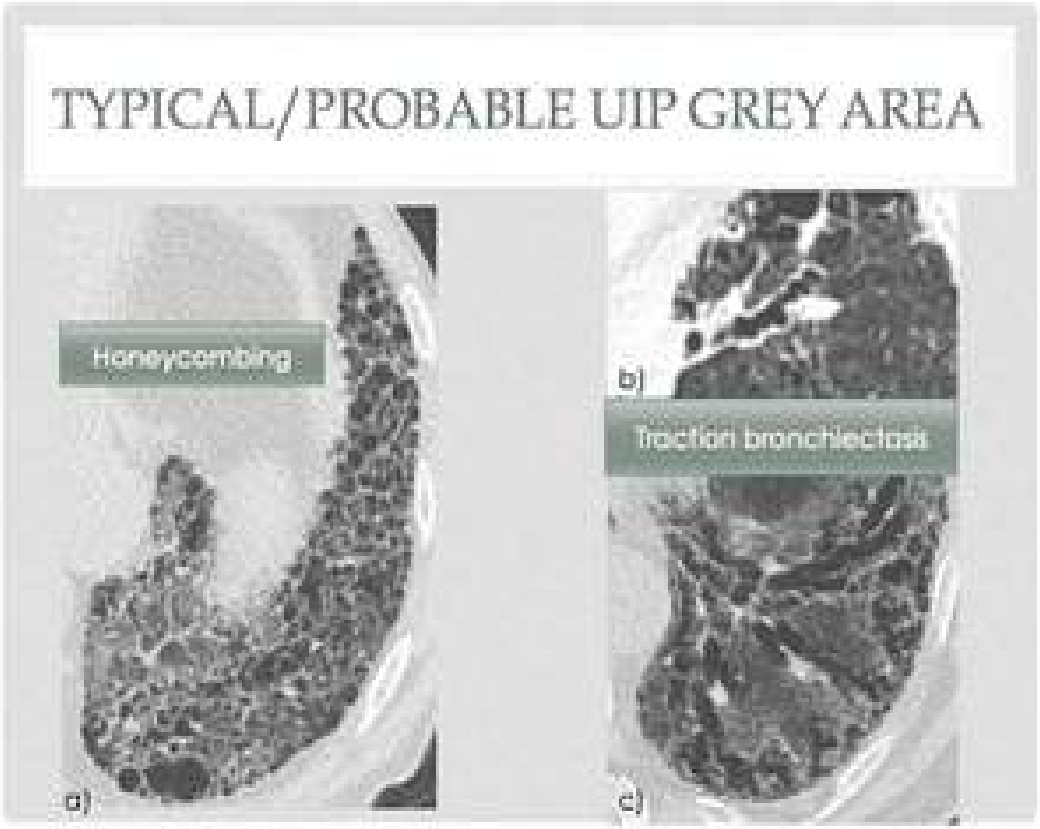
- Alveolar stem-cell exhaustion
- Basal cell dysfunction
- Abnormal extracellular matrix remodelling
- Bronchiolisation
- Honeycomb cyst formation

Aberrant remodelling



TRACTION BRONCHIECTASIS

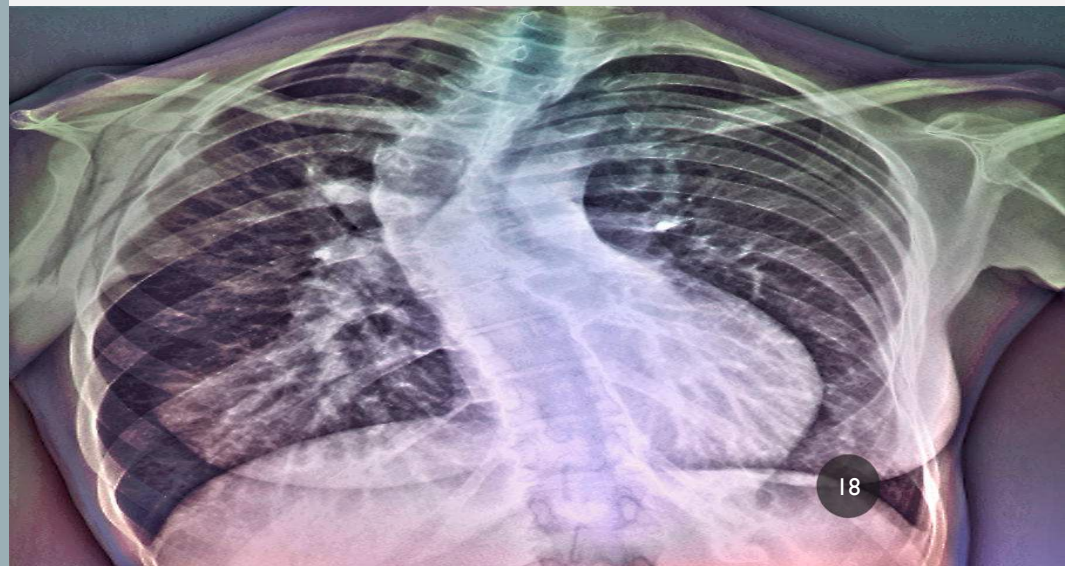
- Occurs when fibrotic tissue, such as that seen in interstitial lung diseases (ILDs) or pulmonary fibrosis, pulls and distorts the surrounding airways.
- The traction or pulling effect leads to the permanent dilation and thickening of the bronchial walls.
- Its presence on HRCT scans has been associated with worse outcomes, including disease progression, increased risk of acute exacerbations, and decreased survival rates. Traction bronchiectasis serves as an indicator of advanced fibrosis and may help guide treatment decisions and prognostic discussions with patients.



Traction bronchiectasis is a radiological finding and does not represent a specific disease entity but rather a consequence of underlying fibrotic lung diseases.

KYPHOSCOLIOSIS

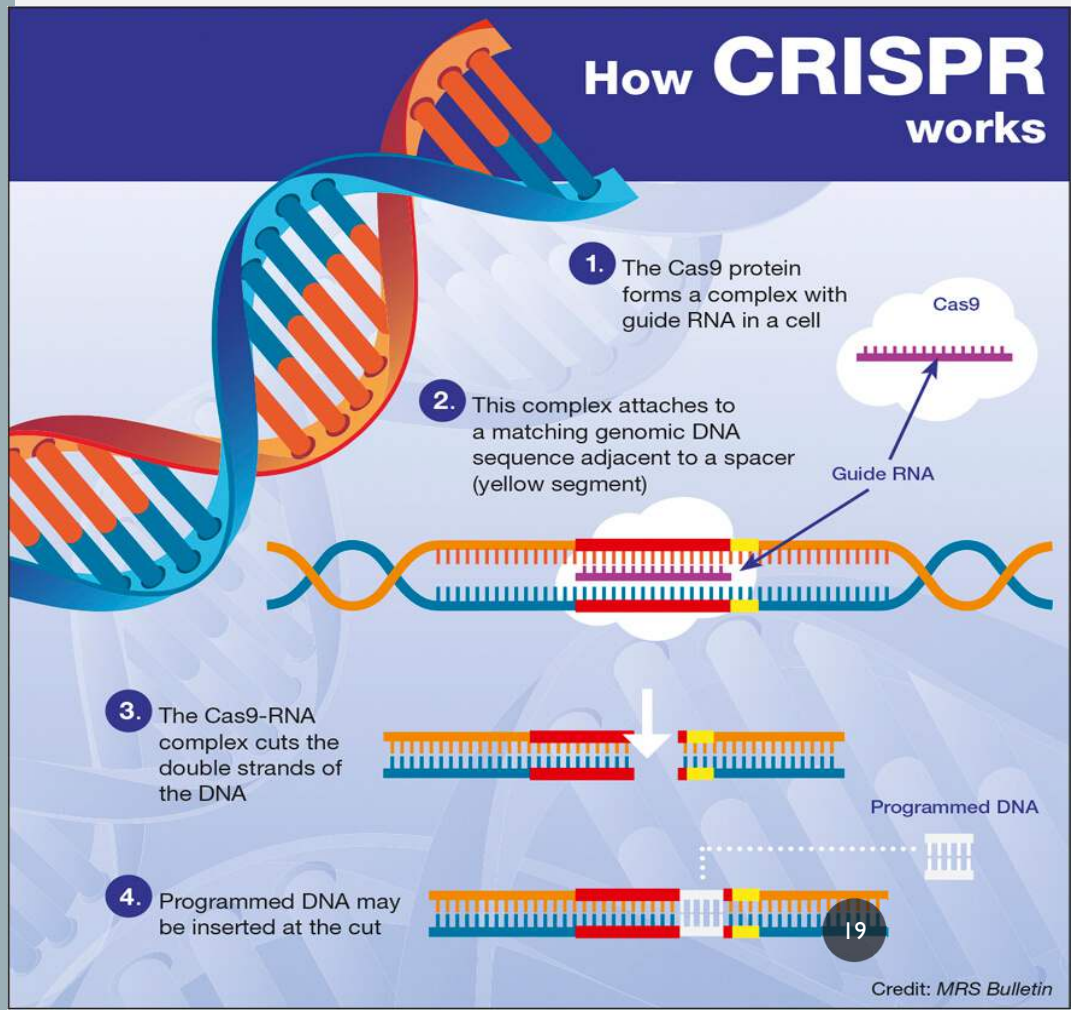
- The abnormal spinal curvature in kyphoscoliosis alters the position and shape of the rib cage, reducing its ability to expand and contract during breathing.
- The abnormal curvature of the spine in kyphoscoliosis can cause uneven distribution of ventilation (airflow) and perfusion (blood flow) within the lungs.
- In progressive kyphoscoliosis, the severity of the spinal curvature and its impact on lung function can worsen over time. This progressive decline in lung function may require ongoing monitoring and interventions to manage respiratory symptoms effectively.
- The impaired clearance of mucus from the airways due to restricted lung function in kyphoscoliosis can predispose individuals to respiratory infections, such as pneumonia and bronchitis. Infections can further exacerbate respiratory symptoms and compromise lung function.
- Treatment options? Surgery, pulmonary rehabilitation, NIV, HFT.



FUTURE OUTLOOK

- Gene modulation/therapy
  - Viral vectors**
  - Non-viral vectors**
  - Nanoparticle* – yea, I said it.
- Phosphodiesterase 4B inhibitor
  - The PDE4B subtype, which has an important role in inflammation and fibrosis, has been identified in the lungs. Preferentially targeting PDE4B has the potential to drive anti-inflammatory and antifibrotic effects via a subsequent increase in cAMP, but with improved tolerability.
- Corrective surgery
- Phrenic pacing or correction

- Why is early detection important
- Prognosis and QOL



CONCLUSION

- Air goes in and out per physics and encounters a lot in its way
- RLD is, well, complicated
- There are over 200 different types of restrictive/interstitial lung disease
- Early Intervention is key in prolonging life and enhancing QOL
- Rates of disease vary among certain groups, and as a whole.
- Intrinsic vs extrinsic
- Inflammation is a key contributor to the process

- Spirometry is handy but lung volumes are best
- CT is the gold standard for diagnosis but there is further testing to really target the problem
- Treatment is exactly that – treatment. It is not curative and is insanely expensive
- Occupational history is also large component
- Traction bronchiectasis may be an indicator for further diagnostics
- The future outlook for RLD is one of microscopic star wars

- FIN